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CASE REPORT

Multiple calcifying fibrous tumors in the small intestine and the mesentery

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inflammatory pseudotumor;
peritoneal tumor;
mesentery tumor

Summary Calcifying fibrous tumor (CFT) is a rare and benign tumor. Histological features are hyalinized fibroblast-poor tissue, infiltrative inflammatory cells mainly composed of lymphocytes or plasma cells, and psammoma bodies or dystrophic calcification. The pathogenesis is unknown. We treated a 30-year-old man who suffered from abdominal pain, nausea and vomiting for 2 days. After X-ray examination, we did not find any ileus, hollow organ perforation or genitourinary tract abnormalities and therefore strongly suspected acute ruptured appendicitis. An appendectomy was therefore performed immediately. Multiple tumors were found during surgery, and pathological examination confirmed the diagnosis of CFT. After surgery, the previous symptoms were relieved and the patient recovered completely. However, another nodule in the greater omentum was found by a computed tomography abdominal scan 4 months later. For CFT with a large tumor and recurrent symptoms, we recommend complete excision as the best treatment. For a case of asymptomatic CFT with a small tumor, long-term follow-up should suffice.

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1. Introduction

Calcifying fibrous tumor (CFT) is an uncommon and benign tumor which was first described by Rosenthal and Abdul-Karim in 1988.¹ Most CFT cases have been reported in children and young adults.²

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CFT has been reported in various sites, such as soft tissues, the mesentery, the gallbladder, the peritoneum, and the pleura.^{1,3-6} However, multiple lesions are rare, and few cases have been reported, especially for mesenteric lesions.⁶⁻⁸ We searched PubMed using the key words "multiple calcifying tumor" and "mesentery," and found only 3 references. We add a new case of a male with multiple lesions in the mesentery, the small intestine and the mesenteric lymph node, and review the published papers on similar cases.

2. Case report

A 30-year-old male had an abdominal pain radiating to the right lower quadrant, accompanied by nausea and vomiting, for 2 days. The patient had had no other medical or systemic disease.

Physical examination, showed abdominal tenderness in the peri-umbilical area and the right lower quadrant with mild rigidity and distension. There was an abnormal WBC of $17,600/\text{mm}^3$. We then checked the patient's X-ray and highly suspected ruptured appendicitis; therefore, an appendectomy was performed immediately (Fig. 1). Since multiple nodules of a tan-white color were found in the small intestine, the mesentery and the mesenteric lymph node during the operation, we decided to perform local excision and a biopsy. The specimens were dissected and consisted of parts of the following: a mesenteric tumor (Specimen A), the small intestine (Specimen B), a mesenteric lymph node (Specimen C) and the appendix (Specimen D). They were all placed in formalin and sent to pathology for definitive diagnosis (Figs. 2A, and 2C, 2B). Specimen A had >10 miliary tumor masses sizing up to $0.8 \times 0.5 \times 0.4$ cm and was attached to the mesenteric surface. Specimen B contained multiple tumor masses measuring up to $1.0 \times 0.4 \times 0.4$ cm. The size of Specimen C was $1.0 \times 0.5 \times 0.4$ cm, and Specimen D was 4.1 cm in length

with no perforation. However, all the sections from Specimens A, B and C showed similar tumor masses which were well-demarcated and bore hypocellular tumors composed of fibroblast-like cells and sporadic inflammatory cells in a highly sclerotic background. Psammoma-like calcifications were easily seen (Fig. 3). There were atypical cells, necrosis or malignancy in any of these specimens. Immunohistochemically, the specimens were negative for CD34, CD117 and desmin but not for vimentin. Based on the above morphological and immunohistochemical results, a diagnosis of CFT was made. Panendoscopy was performed soon after the operation, but no specific finding was observed. Four months later, we followed up with a computed abdominal tomography scan, and a 7 mm nodule in the greater omentum was detected. The patient however denied any discomfort and, as a result, refused further medical investigation.

3. Discussion

CFT is a rare and benign tumor which has distinctive features, including hyalinized fibroblast-poor tissue, infiltrative inflammatory cells mainly composed of lymphocytes or plasma cells, and psammoma bodies or dystrophic calcification. In Table 1, the clinical features of CFT in the mesenteric region are summarized. To date, only five such cases have been reported including our present case. The gender ratio of females to males was 3:2, the ages of the patients ranged from 17 to 41 years, with a mean age of 26 years, and the tumor size varied from 0.8 to 9 cm. Clinical symptoms included abdominal pain, nausea, vomiting, tenderness, rebound pain and abdominal distension. There were two cases, excepting ours, that had experienced acute abdominal pain, nausea and vomiting and were presumed to present appendicitis. One was a 34-year-old male with symptoms in the omentum, and the other a 17-year-old female whose discomforts were in the peritoneal

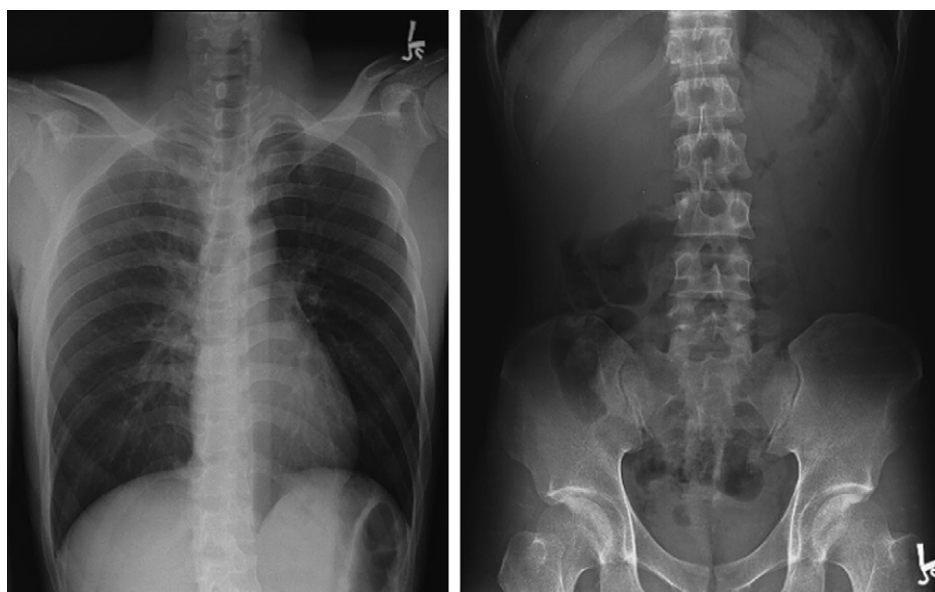


Figure 1 Essentially normal chest x-ray film and non special bowel gas accumulation on plain film.

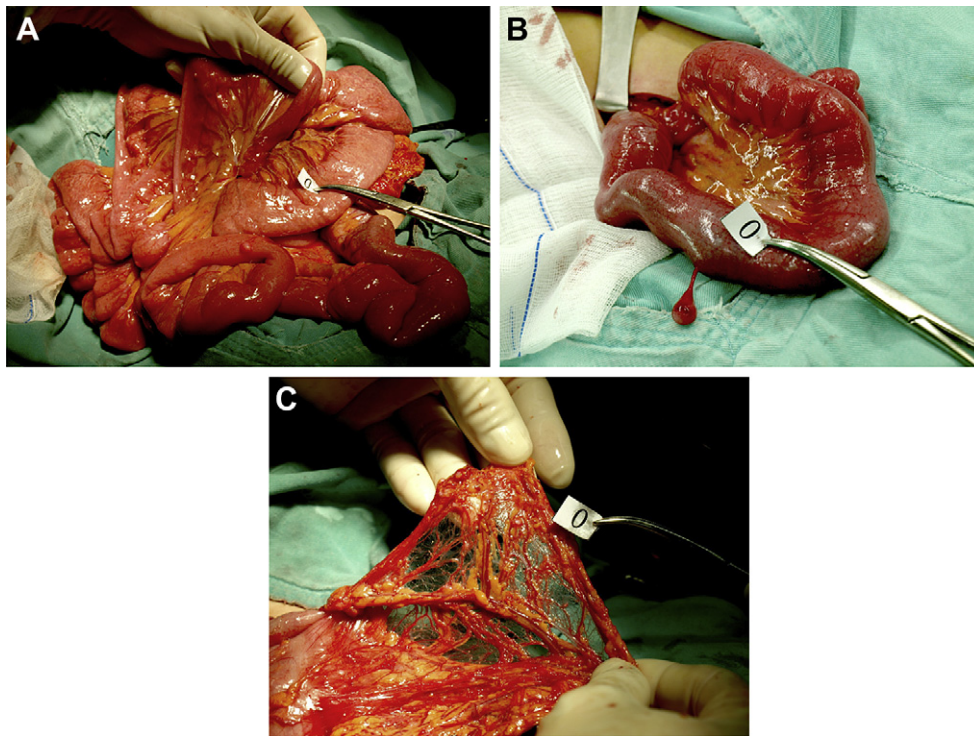


Figure 2 (A) some small mass on mesentery; (B) small pedunculated mass on small bowel wall; (C) multiple small granulation like mass on omentum.

cavity.^{9,10} The symptoms of our case were quite similar to those in the aforementioned two cases, and ruptured appendicitis was therefore diagnosed.

A differential diagnosis of CFT usually includes fibromatosis, nodular fasciitis, inflammatory myofibroblastic tumor (IMT), calcifying aponeurotic fibroma and amyloidoma.⁵ However, CFT differential diagnosis in the GI tract comprises a gastrointestinal stromal tumor (GIST), a desmoid

tumor, IMT, solitary fibrous tumors and a reactive nodular fibrous tumor.⁸ Immunohistochemically, the spindle cells of CFT demonstrate a consistently positive reaction to factor XIIIa and vimentin, but react negatively to smooth muscle actin (SMA), anaplastic lymphoma kinase (ALK), CD34, CD117 and Bcl-2.^{5,8,10–13} For GIST, CD117 is invariably positive. As for IMT, it is positive to SMA, ALK-1 and focally positive to factor XIIIa. Also, the solitary fibrous tumor is positive to CD99 and Bcl-2.^{11–13} Histologically, calcification is a specific feature of CFT, but not seen in the solitary fibrous tumor or nodular fibrous tumor.² Cellularity is great with epithelioid or palisading-vacuolated spindle cells on GIST. Desmoid fibromatosis presents non-multiple nodules, and always lacks calcifications and inflammatory aggregate. IMT is compact dense fibrous-tissues with lymphoid aggregate and no calcifications. In our case, we checked CD34, CD117, desmin and vimentin on the spindle cell, and found it positive only for vimentin. These results were consistent with the immunohistochemical features of CFT.

The recurrence rate of CFT is not high and has been reported to be around 17–30%, mainly in cases of inadequate excision.^{2,3,10} However, there has been no reported recurrence of CFT in the mesentery during the follow-up period. Generally, complete excision is an adequate treatment to avoid recurrence of CFT. However, it may be difficult to clean up minor tumors of <2 mm in size at multiple sites and consequently these tumors will be left untreated, especially when the patient is without any discomfort.⁴ Sleight et al. suggested using repeated CT to monitor the tumor in the pleural or the mediastinal location.¹⁴ In our case, we used gastroscopy to monitor CFT in

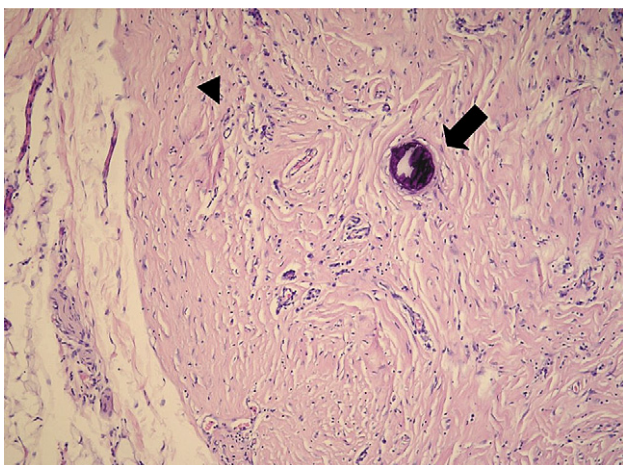


Figure 3 Showing the well-circumscribed, non-encapsulated tumor with psammomatous calcification (arrow) and fibroblast-like cells in a sclerotic background (arrowhead) (H&E stain, 100 \times).

Table 1 Multiple calcifying tumor in the mesenteric region on 5 cases.

Reference	Age/Sex	Symptoms	Location	Size (cm)	Recurrence (Y/N)	F/U (mo)
Chen KT ⁶	19/F	Episodic abdominal pain, recurrent abdominal pain, tenderness	Mesenteric, omentum, small bowel serosa	2	No	228
Liang HH et al. ⁴	17/F	Less episodic abdominal pain, abdominal pain with nausea, tenderness, rebound pain	Mesenteric, small bowel serosal surface	2	No	204
Farah RB et al. ⁷	25/F	Intermittent epigastric cramping, severe abdominal distension, rebound tenderness	Bowel mesentery, small intestine surface	NA	NA	18
Present case	41/M	Abdominal pain, nausea and vomiting	Pelvic, mesocolon, mesorectum, omentum, mesentery	9 × 5.5 × 5	NA	6
	30/M	Abdominal pain, nausea and vomiting	Mesenteric, small intestine, mesenteric lymph node	0.8 × 0.5 × 0.4	No	4

F = female; M = male; NA = not available; F/U = follow up; Y = yes; N = no.

the GI tract, because endoscopy is more efficient and provides clearer demonstration than CT for this purpose.

In summary, we report a case of acute abdominal pain which was suspected to be due to ruptured appendicitis. Nodules at multiple sites were discovered during the operation. Histopathological features of CFT can provide an accurate diagnosis when immunohistochemical stain is not discernable or even confusing. Since there are three reference cases of CFT that were presumed to present appendicitis prior to operation, we suggest when diagnosing any acute abdominal pain of unknown causes or similar pain lasting for several years, the possibility of CFT should be considered. When the tumor is too small and the patient is without any discomfort, usual diagnostic instruments normally cannot identify the lesion, thus, leaving multiple CFTs undetected, should they exist. Because of the low recurrence rate of CFT and the elapse of many years for such a recurrence to take place, we suggest that a long-term follow-up be undertaken, and any symptomatic recurrence should be treated by re-excision.

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